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## An unusual presentation of EATL type 1: Emergency surgery due to life-threatening gastrointestinal bleeding

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## ABSTRACT

**INTRODUCTION:** Enteropathy-associated T-cell lymphoma (EATL) is a very rare malignancy. Reasons for hospital admission are variable.**PRESENTATION OF CASE:** 76 years old man admitted to emergency service with sudden and massive obscure gastrointestinal bleeding. There was no complaints in his history. After initial evaluation, emergency laparotomy had to be done. Bleeding lesion in proximal jejunum was resected. Histopathologically, the muscularis propria had abundant atypical lymphoid infiltrate in diffuse pattern. Atypical lymphoid cells expressed CD3 and CD30. The jejunal mucosa adjacent to the tumor showed effacement of normal villous architecture.**DISCUSSION:** EATL is known to cause anemia as a result of chronic bleeding. However in this case, the bleeding was abundant, irreplaceable and requiring emergency surgery. To our knowledge it is not reported previously.**CONCLUSION:** A sudden and massive gastrointestinal bleeding can be the first and unique sign of EATL.

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## 1. Introduction

Enteropathy-associated T-cell lymphoma (EATL) is an intestinal tumor arise from intraepithelial T-cells.<sup>1</sup> It is a very rare disease with the annual incidence rate is 0.5 to 1 per million.<sup>2</sup>

EATL is strongly associated with celiac disease (CD).<sup>1,3–6</sup> Although CD is a long standing disease, EATL is more often preceded by a short history of adult CD diagnosed after 50 years of age. In some cases, there is no history of CD, but histopathologic features of it are found in the tissue surrounding the tumor after resection.<sup>1,7</sup> In 2008, two types of EATL are introduced into the World Health Organization (WHO) classification of tumors of hematopoietic and lymphoid tissues.<sup>8</sup> The distinction between two types is made according to morphologic and genetic features. Type 1 EATL is more frequently associated with celiac disease.<sup>1,7–10</sup>

Non-specific signs and symptoms can be seen in patients with EATL. Sometimes occult or overt gastrointestinal bleeding or bowel perforation can be reason of hospital admission. Gastrointestinal bleeding seen in patients with lymphoma is usually insidious and

it is manifested by iron deficiency anemia.<sup>1,10,11</sup> To our knowledge, in these patients, sudden and massive gastrointestinal hemorrhage has not reported previously. In this article, we report the very unusual case of EATL which manifested by sudden and life-threatening massive gastrointestinal bleeding, without previously known CD.

## 2. Case report

A 76-years-old male admitted to emergency service with sudden and severe gastrointestinal bleeding and hypovolemic shock. There were massive hematochezia and hematemesis. There is no history of celiac disease in his/family history. Systolic arterial blood pressure was 60 mmHg, diastolic was unmeasurable. The patient was monitored and resuscitation were started immediately. Crystalloids and erythrocyte suspensions were given intravenously. After the initial resuscitation hemodynamic stability was provided and then the patient transferred to Intensive Care Unit (ICU). The hemoglobin value increased from 4 mg/100 mL to 8 mg/100 mL. Emergency upper and lower gastrointestinal endoscopy revealed no lesion or active bleeding. There was blood clots in the colon and stomach. A short time later, second severe bleeding was observed in ICU. Despite the resuscitation and transfusions, the patient re-entered the hypovolemic shock. We decided to emergency laparotomy in doubt.

In exploration there was an adhesion between the transverse mesocolon and the jejunum just after the ligament of Trietz. We divided the adhesion. There was a perforated lesion that had been bleeding, on antimesenteric surface of jejunum (Fig. 1). Perforated

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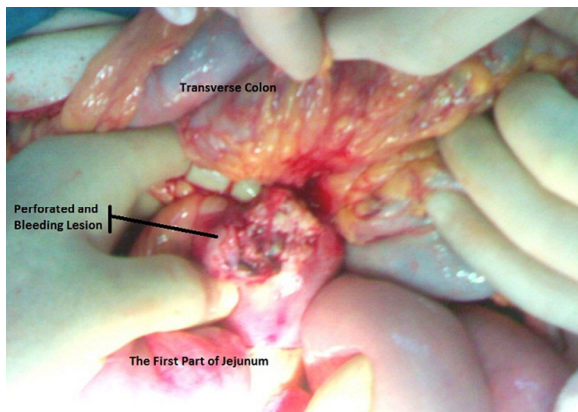


Fig. 1. Perforated and actively bleeding tumor on jejunum.

area was attached to adjacent intestine so that there was no blood in peritoneal cavity. We resected the lesion with margins of a few centimeters of healthy jejunum in both sides. End-to-end anastomosis was performed.

In histopathological examination, macroscopically, there were traces of bleeding and almost complete necrosis. Microscopically,

intestinal mucosa showed extensive ulceration and necrosis. Extensive necrosis made the diagnosis very hard. But the following findings led us to the diagnosis of EATL type 1. The muscularis propria had abundant atypical lymphoid infiltrate in diffuse pattern (Fig. 2a). Neoplastic lymphocytes were medium to large sized cells with irregular nuclear contours and hyperchromatic nuclei. Some large cells with vesicular nuclei had prominent nucleoli (Fig. 2c). The jejunal mucosa adjacent to the tumor showed effacement of normal villous architecture (villous atrophy, crypt hyperplasia, intraepithelial lymphocytosis, pyloric metaplasia) (Fig. 3a and b). Immunohistochemically, atypical lymphoid cells positive for CD3 (Fig. 2b) and CD30 (Fig. 2d). They were immunonegative for CD56, CD4, CD8, CD20, ALK1. The background population was composed of reactive B and T lymphocytes. The intraepithelial lymphocytes in the adjacent mucosa were positive for CD3 and negative for CD4 and CD8.

The patient was discharged after surgical healing and referred to hematology clinic in eighth day postoperatively. The patient received combined chemotherapy regimen composed of bleomycin, doxorubicin, cyclophosphamide, vincristine, and prednisone. Since then he has been followed up by 18F-fluorodeoxyglucose positron emission tomography. The patient is alive in twenty-fourth month after diagnosis, there were no

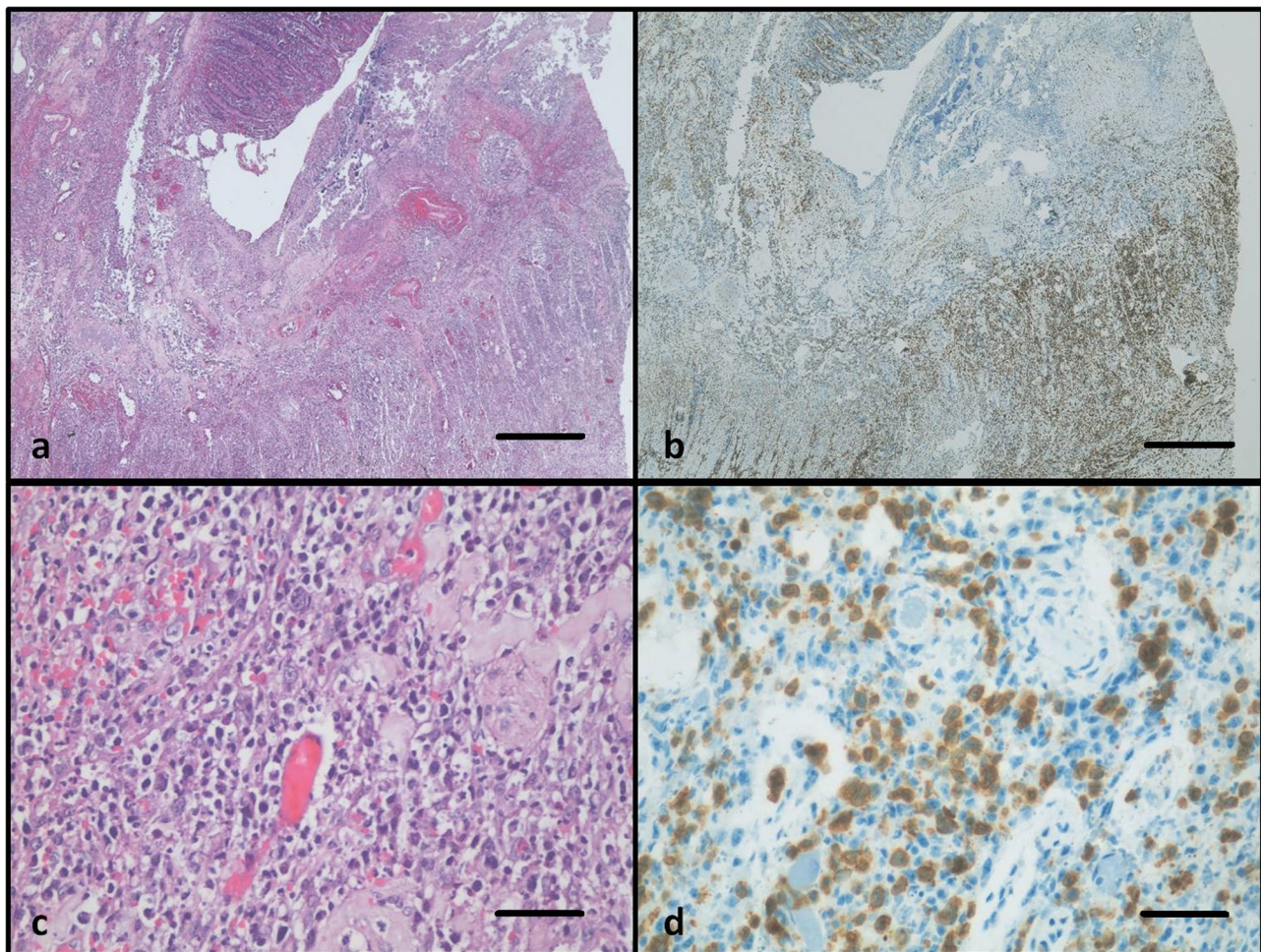
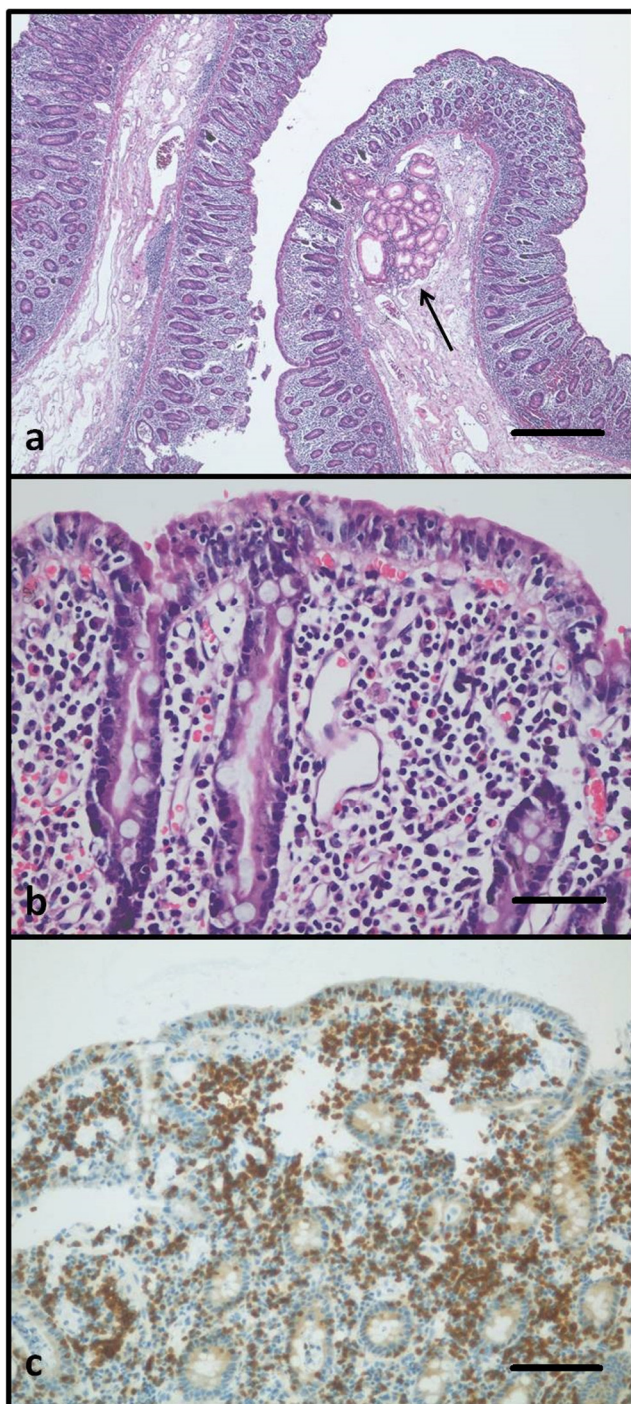


Fig. 2. (a) Extensive mucosal ulceration and diffuse lymphoid infiltration in submucosal and muscular layer of jejunal wall (hematoxylin-eosin, original magnification 20 $\times$ ). (b) Immunohistochemically, tumor cells are reactive for CD3 (immunoperoxidase, original magnification 20 $\times$ ). (c) At high power magnification, variably sized atypical pleomorphic lymphocytes within a necrotic background (hematoxylin-eosin, original magnification 400 $\times$ ). (d) The neoplastic cells commonly express CD30 (immunoperoxidase, original magnification 400 $\times$ ). Scale bar = 100  $\mu$ m.





**Fig. 3.** (a) At low power magnification, adjacent mucosa shows blunting of the villous architecture, crypt hyperplasia, and pyloric metaplasia (arrow) (hematoxylin-eosin, original magnification 40 $\times$ ). (b) At high magnification, adjacent mucosa shows numerous intraepithelial lymphocytes accompanied by a predominantly plasmacytic inflammatory infiltrate with scattered eosinophils in the lamina propria (hematoxylin-eosin, original magnification 200 $\times$ ). (c) Intraepithelial lymphocytes are positive for CD3 (immunoperoxidase, original magnification 100 $\times$ ). Scale bar = 100  $\mu$ m.

findings suggesting recurrence or metastasis. He continues to strict gluten-free diet.

### 3. Discussion

According to the WHO classification of neoplasms of the hematopoietic and lymphoid tissues, EATL is recognized that a

variety of T-cell lymphomas can present with intestinal involvement, but not all are associated with CD.<sup>8</sup> There are two types of EATL, each has different genetic, morphologic and immunologic features. Type 1 EATL consists of medium-sized to large or pleomorphic cells and it is associated with CD and the HLA-DQ2 haplotype. The cells in type 1 EATL can be immunopositive for CD3 and CD30. Type 2 EATL consists of monomorphic small to medium sized cells. Cells in type 2 EATL are frequently express CD56.<sup>1,7–10,12</sup>

EATL occurs predominantly in sixth decade of life and is slightly more common in men. It is commonly localized to small intestine especially in jejunum.<sup>1,10,12,13</sup> EATL tumors are often multifocal and can present with weight loss, abdominal pain, change in bowel habit, vomiting, diarrhea or even acute abdomen due to ulceration, obstruction or perforation.<sup>13,14</sup> Occult or overt bleeding from gastrointestinal tract can also be seen in EATL.<sup>1,10,11</sup> To our knowledge this is the first case of sudden and life-threatening gastrointestinal bleeding in EATL cases.

It is difficult to diagnose the EATL. Patients with EATL need a thorough workup including video capsule enteroscopy, double balloon enteroscopy, computed tomography, 18F-fluorodeoxyglucose positron emission tomography, magnetic resonance enteroclysis.<sup>1</sup> In present case, the patient has never been any gastrointestinal complaints. In addition, when we consider the urgency of the case, there was no chance to evaluate the patient preoperatively.

The source of gastrointestinal bleeding is commonly identified by conventional upper or lower GI endoscopy. But approximately 5% of all cases remain undiagnosed.<sup>15</sup> Obscure gastrointestinal bleeding (OGIB) is defined as bleeding of unknown origin that persists or recurs after initial endoscopic evaluation. It can be overt or occult.<sup>16</sup> According to American Society for Gastrointestinal Endoscopy (ASGE), in the cases of active overt OGIB, after initial endoscopic evaluation, the preferred intervention is angiography.<sup>17</sup> But in our case, we encountered massive and recurrent gastrointestinal bleeding that was not appropriate to algorithm of ASGE. After endoscopic evaluation in compliance with the algorithm for obscure gastrointestinal bleeding we had to make an emergency laparotomy. If we did not find the source of bleeding, intraoperative enteroscopy would be a good option.<sup>18</sup>

EATL is a serious disorder that require long-term hemathologic treatment. However, we did not emphasize the hemathologic treatment of the patient here because it is not the subject of this article.

In conclusion, EATL is a rare but very important disease with poor prognosis and presentation of the disease is variable. In addition to what we know up to now, this case show us a sudden and massive gastrointestinal bleeding can be the first and unique sign of EATL.

### Conflict of interest statement

None.

### Funding

None.

### Ethical approval

We declare that we have obtained written consent from the patient for surgical intervention.

### Author contributions

Erdem Kinaci worked in study design, data collection and writing.

Mehmet Emin Gunes worked in data collection and writing.

Gulben Erdem Huq worked in histopathological examination and data collection.

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